An Unusual Variant of Confluent and Reticulated Papillomatosis Masquerading as Tinea Versicolor

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Background: Confluent and reticulated papillomatosis (CARP), also known as Gougerot-Carteaud syndrome, is a rare disorder. It usually presents as hyperkeratotic brown papules that coalesce into plaques with a reticulated periphery on the central trunk of young adults. Confluent and reticulated papillomatosis is most often clinically confused with tinea versicolor and usually does not respond to therapy with antifungals. Minocycline is the treatment of choice.

Observations: Four cases of CARP with the unusual presentation of hypopigmented lesions masquerading as tinea versicolor in dark-skinned (Fitzpatrick skin types IV-V) patients are presented. All cases exhibited characteristic features of CARP on biopsy results and responded to minocycline of several months’ duration. Two of the cases were also treated with adjuvant topical tazarotene.

Conclusions: The hypopigmented variant of CARP in dark-skinned patients makes the clinical differentiation from tinea versicolor extremely challenging. Physicians encountering darkly pigmented individuals with hypopigmented plaques unresponsive to antifungals should have a high clinical suspicion for the hypopigmented variant of CARP.


Confluent and reticulated papillomatosis (CARP) (Gougerot-Carteaud syndrome) is a rare disorder first characterized by Gougerot and Carteaud in 1927. The clinical presentation of CARP is that of hyperkeratotic or verrucous brown papules that coalesce into plaques with a reticulated periphery. This condition usually occurs on the central trunk in young adults. Histopathologic examination reveals hyperkeratosis, squat papillomatosis, and acanthosis. Occasionally, a perivascular lymphocytic infiltrate is seen in the superficial dermis.

Confluent and reticulated papillomatosis is most often clinically mistaken for tinea versicolor and usually does not respond to therapy with antifungals. A variety of topical and oral therapies exist, but minocycline is the current treatment of choice. Despite therapy, recurrence is possible.

REPORT OF CASES

CASE 1

A 36-year-old African American woman with Fitzpatrick skin type V presented with a pruritic eruption on the central aspect of the chest, abdomen, and back that she had had for 4 months. The patient had been using selenium sulfide shampoo, 2.5%, and topical ketoconazole without relief. On examination, the lesions were described as scaling plaques that were hypopigmented in a reticulated pattern. For the next 2 years, the patient was treated with selenium sulfide shampoo, 2.5%, 2 courses of ketoconazole, 400 mg/d, for 5 days; and 1 course of fluconazole, 200 mg, every 3 days for 1 week. The lesions continued to be pruritic and spread to her face and neck.

Potassium hydroxide preparations produced negative results. Biopsy results revealed hyperkeratosis, low papillomatosis, acanthosis, basal layer pigmentation, and a sparse perivascular lymphocytic infiltrate in the superficial dermis. A period acid–Schiff stain yielded negative results for fungal and yeast organisms.

The patient was diagnosed as having CARP. She was treated with minocycline, 100 mg/d, for 3 months and then tapered to every other day for 2 months, after which her lesions cleared. The patient remained free of lesions at 3 months of follow-up.

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A 15-year-old Latino boy with Fitzpatrick skin type IV presented with a rash on his chest and back. He had been diagnosed as having tinea versicolor by an outside dermatologist and treated with econazole nitrate cream without improvement and with progression of his lesions. On examination, he was found to have scaling hypopigmented macules that coalesced into large plaques on the central trunk. He was treated with selenium sulfide shampoo, 2.5%, daily. He also was prescribed ketoconazole, 400 mg, once, which was followed by an hour of sweating; this treatment was repeated in 1 week.

The patient was lost to follow-up and returned to the clinic 3 years later. He reported persistent lesions. Examination revealed hypopigmented macules and patches on his neck, lower abdomen, and lower back. Biopsy results revealed mild acanthosis and papillomatosis of the epidermis. In the dermis, there was a mild to sparse perivascular lymphohistiocytic infiltrate. A periodic acid–Schiff stain result was negative for fungal elements, although rare spores were noted.

The patient was diagnosed as having CARP and treated with minocycline, 100 mg, twice daily. After 3 months of treatment, the patient reported minimal response to minocycline. Daily application of tazarotene cream, 0.05%, was added as adjuvant therapy, and the patient continued with both medications for another 2 months. At follow-up, the patient reported vast improvement. Minocycline therapy was discontinued, but daily use of tazarotene cream, 0.05%, was continued for maintenance. The patient continued using tazarotene alone and remained free of rash after 3 months.

CASE 3

A 17-year-old Latina girl with Fitzpatrick skin type IV presented for a second opinion regarding a 3-year history of a mildly pruritic rash on her chest, abdomen, and back. She had been treated with selenium sulfide shampoo, 2.5%, for 2 months without improvement. On examination, she had multiple, slightly scaly, hypopigmented macules and papules on her neck, central chest, shoulders, and upper back. Potassium hydroxide preparation did not reveal hyphae or spores. A biopsy performed by an outside dermatologist revealed only a few melanophages in the papillary dermis. The patient refused another biopsy, opting instead for treatment.

The patient was diagnosed as having CARP and was prescribed minocycline, 100 mg/d. After 2 months of therapy, the lesions improved; the minocycline dosage was decreased to 100 mg every other day for 1 month. However, on tapering, she began to relapse. The minocycline dosage was changed to 50 mg/d and adjuvant therapy with topical tazarotene cream, 0.1%, daily was added, which the patient used for 2 weeks. The patient remained free of rash while not receiving any therapy at the 6-month follow-up.

CASE 4

A 23-year-old African American woman with Fitzpatrick skin type V presented with a light-colored rash on her chest and trunk that she had had for several months. Lesions were described as faint hypopigmented macules that coalesced into patches. Biopsy revealed papillomatosis, hyperkeratosis, and mild acanthosis with mild perivascular dermal inflammation, consistent with CARP. Treatment was initiated with minocycline, 100 mg, twice daily and ammonium lactate cream, 12%, twice daily. After 6 weeks of oral and topical therapy, the patient's lesions resolved.

Four months later, the patient returned with a recurrence of her lesions. She was again prescribed minocycline, 100 mg, twice daily for 2 months. Two years later, she again returned with a recurrence, stating that her lesions had been cleared with prior therapy. She resumed using minocycline at a dose of 100 mg/d. She was again lost to follow-up.

COMMENT

Four cases of an unusually hypopigmented variant of CARP masquerading as tinea versicolor in dark-skinned (Fitzpatrick skin types IV-V) patients are presented. The diagnostic criteria for CARP proposed by Davis et al require (1) clinical findings of scaling brown macules and patches, some reticulated and papillomatous; (2) location on the upper trunk and neck; (3) fungal staining of scale negative for spores and hyphae; (4) lack of response to antifungals; and (5) excellent response to minocycline. The aforementioned patients met these criteria with the exception of the color of the lesions, thus expanding the clinical presentation of CARP to include hypopigmented lesions. The uncharacteristic light color of CARP lesions in our patients delayed a correct diagnosis by a mean of 2.25 years. The hypopigmented eruption was misdiagnosed as tinea versicolor, although the differential diagnosis of this unusual variant of CARP includes other entities as well (Table). The
only other publication focusing on the color of CARP lesions is a case series of 3 white teenagers presenting with persistently red lesions described as “nonpigmenting” CARP. It is likely that the present case series of hypopigmented CARP involves dark-skinned patients only because the hypopigmentation is more noticeable in these patients compared with white patients, although it remains to be seen whether hypopigmented CARP presents in patients with Fitzpatrick skin types III or less.

The pathogenesis of CARP remains unknown, but most studies favor defective hyperkeratination. Electron microscopic examination has shown increased numbers of transitional cells and foci of keratin 16, a marker of hyperproliferation. Although most authorities differentiate CARP from tinea versicolor, a report of 3 siblings simultaneously suggests a possible pathogenic link or genetic predisposition. Supporting studies have hypothesized an inherited tendency to a hyperproliferative response to colonization by Malassezia furfur, the fungus that causes tinea versicolor. This association may further explain the similar clinical appearance of the 2 entities, in particular the hypopigmented lesions described in our patients. However, any association between CARP and tinea versicolor remains speculative at this time.

The pathophysiology of the hypopigmentation of the unusual variant of CARP is even less clear because any of the basic processes of melanin pigmentation may be defective. A Fontana-Masson stain for melanin was performed, which revealed decreased melanin in lesional compared with nonlesional skin (Figure 2), yet a Mart-1 stain revealed a normal distribution of melanocytes in both lesional and nonlesional skin. Such findings suggest that decreased melanin synthesis may cause the hypopigmentation seen in the unusual variant of CARP, but the exact mechanism for this is unknown.

The current treatment of choice for CARP is oral minocycline, 50 to 100 mg, twice daily for 6 weeks. It is unclear how and why minocycline works, but its anti-inflammatory properties may be responsible. Similar to patients with typical CARP, patients with hypopigmented CARP responded well to minocycline. However, these patients required 2 to 5 months of minocycline, 100 mg once or twice daily. This longer duration of treatment may be caused by delayed diagnosis or a higher rate of relapse in patients with hypopigmented CARP. Because postinflammatory hypopigmentation from inflammatory conditions, such as tinea versicolor, takes approximately 6 months to clear, the fact that the patients vastly improved or were free of rash in less than 6 months with minocycline therapy supports a diagnosis of hypopigmented CARP and not postinflammatory hypopigmentation from previously treated tinea versicolor.

In addition to oral antibiotics, topical retinoids are an effective treatment for CARP and were used as adjuvant therapy in recalcitrant cases of hypopigmented CARP. Bowman and Davis described an 11-year-old African American girl successfully treated with tazarotene gel, 0.1%, twice daily for 2 months, with subsequent intermittent use for local recurrences. Similarly, Schwartzberg and Schwartzberg presented 3 cases of CARP that responded to topical tretinoin gel, 0.025% or 0.1%, daily. In 1 of the patients, the rash almost completely cleared after 10 weeks of treatment. Likewise, 2 of our patients with hypopigmented CARP had a good response to tazarotene cream, 0.05% or 0.1%, used daily as an adjunct to minocycline.

Table. Differential Diagnosis of Hypopigmented CARP

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Characteristic Feature(s)</th>
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<tr>
<td>Hypopigmented CARP</td>
<td>Potassium hydroxide scraping and PAS stain negative for fungal elements</td>
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<tr>
<td>Tinea versicolor</td>
<td>Potassium hydroxide scraping and PAS stain positive for fungal elements, Responds to antifungal therapy</td>
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<tr>
<td>Postinflammatory hypopigmentation</td>
<td>History of inflammatory lesions, Possible prior use of topical corticosteroids, which can contribute to hypomelanosis</td>
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<tr>
<td>Pityriasis alba</td>
<td>Responds to gentle bathing habits and topical corticosteroids</td>
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<tr>
<td>Mycosis fungoides</td>
<td>Responds to gentle bathing habits and topical corticosteroids</td>
</tr>
<tr>
<td>Trichrome vitiligo</td>
<td>Perifollicular regrowth</td>
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<tr>
<td>Progressive macular hypomelanosis</td>
<td>May be associated with Propionibacterium acnes</td>
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<tr>
<td>Sarcoidosis</td>
<td>Granulomatous inflammation on biopsy</td>
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</tbody>
</table>

Abbreviations: CARP, confluent and reticulated papillomatosis; PAS, periodic acid–Schiff.

Figure 2. Decreased melanin in lesional skin. Comparison of lesional skin (A) and nonlesional skin (B). A, Architectural features consistent with confluent and reticulated papillomatosis include hyperkeratosis, low papillomatosis, acanthosis, and a sparse perivascular, lymphocytic infiltrate in the superficial dermis (Fontana-Masson, original magnification ×10).
Confluent and reticulated papillomatosis is a rare disorder that is most often clinically confused with tinea versicolor, even when CARP presents in a typical fashion. The current report of an unusual variant of the hypopigmented variant of CARP in dark-skinned patients underscores the importance of keeping a high clinical suspicion for CARP in patients who present with lesions that resemble tinea versicolor, in particular in darkly pigmented individuals in whom antifungal therapy has failed. Hypopigmented CARP responds to minocycline but appears to require several months of treatment and adjuvant topical tazarotene in recalcitrant cases.

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REFERENCES